

# Paraneoplastic Pseudoachalasia in a Patient with Metastatic Salivary Gland Tumor

Venkatesh Vaithiyam<sup>1\*</sup>, Siddharth Srivastava<sup>2</sup>, Surbhi Goyal<sup>3</sup>, Sanjeev Sachdeva<sup>4</sup>

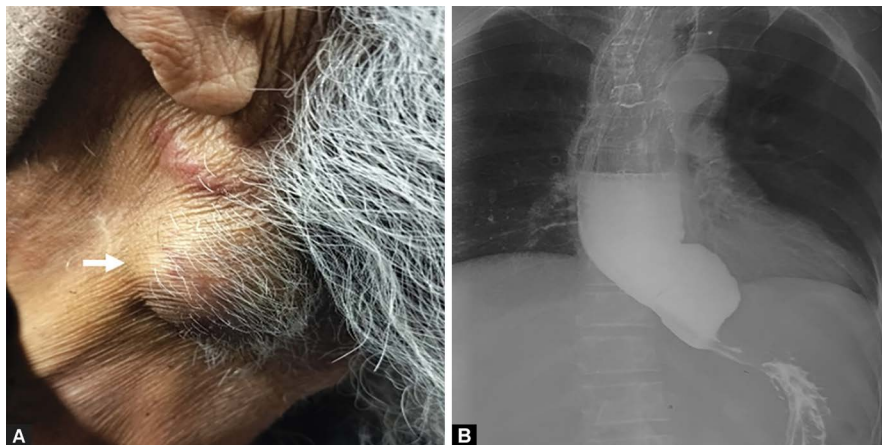
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**A** 73-year-old man presented with a 6-month history of dysphagia and regurgitation of food particles. He also

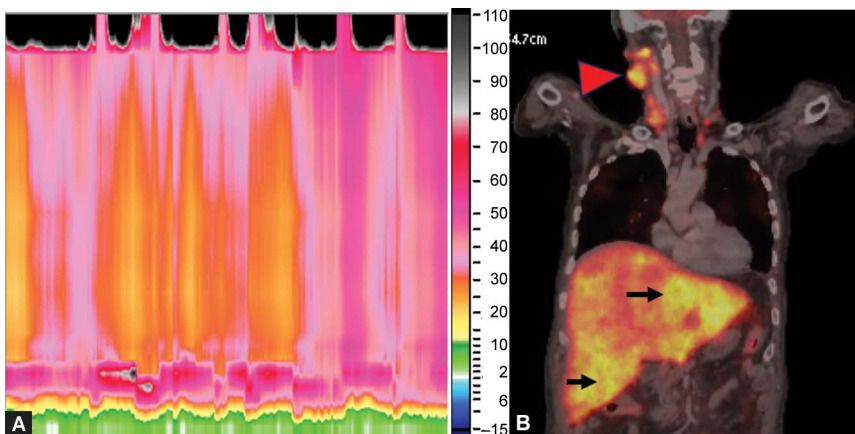
complained of a hard swelling in the right cheek, jaundice, and a 10 kg weight loss over the past month. Examination showed

pallor, icterus, nontender, hard right salivary gland swelling (Fig. 1A), and severe cachexia (body mass index 15.2 kg/m<sup>2</sup>). Abdominal ultrasonography revealed multiple hypoechoic space-occupying lesions (SOL) in the liver, suggesting hepatic metastasis. Upper GI endoscopy, barium swallow (Fig. 1B), and high-resolution manometry (Fig. 2A) showed features of achalasia cardia. 18-FDG positron emission computed tomography confirmed a right salivary gland tumor with lymph node, skeletal, and hepatic metastases (Fig. 2B). Cytopathology of the salivary gland mass and liver SOL confirmed metastatic salivary gland tumor (Figs 3A to D). With the presence of a metastatic tumor and clinical, radiological, endoscopic, and manometric patterns resembling achalasia, the possibility of paraneoplastic pseudoachalasia was kept. Due to poor performance status, endoscopic pneumatic dilation was performed; however, he developed hospital-acquired pneumonia and sepsis and died of progressive respiratory failure.

Pseudoachalasia refers to disorders that clinically, manometrically, and radiographically resemble primary achalasia and are seen in various malignant and nonmalignant conditions.<sup>1</sup> Malignancy-associated pseudoachalasia



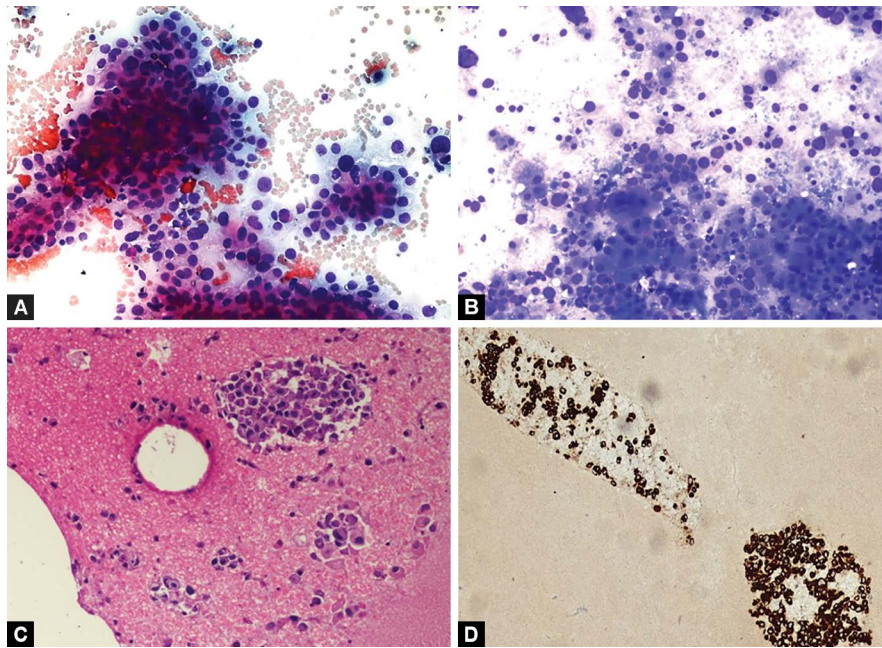
**Figs 1A and B:** (A) Clinical image showing a hard right parotid gland swelling (white arrow); (B) Barium swallow showing dilated esophagus with bird-beak appearance



**Figs 2A and B:** (A) High-resolution manometry showing pan-esophageal pressurization and elevated integrated relaxation pressure; (B) 18-FDG positron emission computed tomography showing a heterogeneously enhancing ill-defined mass lesion in the right parotid region infiltrating the parotid gland (red arrowhead; SUVmax 8.71), suggestive of right parotid gland malignant neoplasm, and FDG-avid extensive hypodense heterogeneously enhancing lesions completely occupying the liver parenchyma (SUVmax 8.12), suggestive of bilobar hepatic metastasis (black arrow)

<sup>1</sup>Assistant Professor; <sup>2,4</sup>Professor, Department of Gastroenterology; <sup>3</sup>Professor, Department of Pathology, Govind Ballabh Pant Institute of Post Graduate Medical Education and Research, New Delhi, India; \*Corresponding Author

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**Figs 3A to D:** FNA from salivary gland: (A) Cytosmears showing tumor cells that were cuboidal to plasmacytoid in appearance, with eccentrically placed enlarged nuclei, moderate eosinophilic cytoplasm, coarse chromatin, and 1–2 prominent nucleoli. Significant cytological atypia and nuclear overcrowding with a few bizarre pleomorphic tumor cells were noted (Papanicolaou  $\times 200$ ); (B) USG-guided FNA from liver SOL: Cytosmears showing a moderate degree of pleomorphism and cytological atypia in the form of a high N:C ratio, nuclear hyperchromasia with coarse chromatin, and 1–2 prominent nucleoli. Many bizarre tumor giant cells were also seen. A few benign reactive hepatocytes were noted in the background (MGG  $\times 200$ ); (C) Cell block showing tumor cells arranged in sheets as well as singly scattered. Individual cells show moderate pleomorphism with a high N:C ratio and nuclear enlargement. Cells are cuboidal with a moderate amount of cytoplasm and have enlarged hyperchromatic nuclei and 1–2 prominent nucleoli (H&E  $\times 100$ ); (D) On IHC, tumor cells were positive for CK7, suggestive of metastatic salivary gland tumor

can be a paraneoplastic syndrome with isolated esophageal involvement or part of a generalized gastrointestinal motility disorder.<sup>2</sup> The pathophysiology involves autoantibody-induced (type 1 antineuronal nuclear autoantibody) destruction of the esophageal myenteric plexus. Treatment includes management of the underlying malignancy and therapies to lower the lower esophageal sphincter pressure.

### INFORMED CONSENT

Informed consent was obtained from the patient's son for the publication of his information and imaging.

### ORCID

Venkatesh Vaithiyam <https://orcid.org/0000-0002-0713-5501>

Siddharth Srivastava <https://orcid.org/0000-0001-7550-0358>

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